RESEARCH LETTER

Tetralogy of Fallot with coronary artery to pulmonary artery fistula

Running head: Tetralogy of Fallot -coronary to artery fistula

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WHAT'S ALREADY KNOWN ABOUT THIS TOPIC?

- Coronary artery fistulas (CAF) are rare anomalies with an incidence of 0.002 % in the population.

WHAT DOES THIS STUDY ADD?

- The association of tetralogy of Fallot, CAF and agenesis of the ductus arteriosus has not been reported
- The discrepancy between the narrow pulmonary annulus with thickened pulmonary valve and the large pulmonary trunk can be caused by extra blood supply through the large fistula.

We report a right coronary artery to pulmonary artery fistula as a rare congenital malformation in a patient with tetralogy of Fallot and absent ductus arteriosus.

A healthy 33-year-old woman, gravida 3, para 1, was referred at 18+4 weeks with suspicion of a fetal cardiac malformation. Her brother died neonatally from a transposition of the great vessels.

Ultrasound examination in this pregnancy revealed a tetralogy of Fallot (TOF) with a right-sided aorta and a narrow pulmonary artery annulus with only mild antegrade flow, while the pulmonary trunk was well developed (Figure 1). No extracardiac anomalies were seen. An amniocentesis showed a normal male CNV-profile.

The couple decided to terminate the pregnancy at 23+5 weeks of gestation. The autopsy showed a TOF with a right-sided aortic arch and agenesis of the ductus arteriosus. Remarkable is the wide origo of the right coronary artery with a larger artery branching from this right coronary artery and passing ventrally of the pulmonary truncus and draining above the left anterior cusp into the lumen of the pulmonary truncus (Figure 2). The left coronary artery was normal and no extracardiac anomalies were found.

Congenital coronary artery fistulas (CAF) are characterized by a normal aortic origin of the coronary artery involved but with a fistulous communication with the atria, the ventricles or with the pulmonary artery, coronary sinus or the superior vena cava. Coronary artery fistulas are rare anomalies encountered in 0.2% of angiographic series. The incidence in the overall population is estimated 0.002 %. The right coronary artery is most often involved in clinically detectable CAF. Fistulas most frequently involve the right ventricle, closely followed, in terms of incidence, by drainage into the right atrium or pulmonary artery. Left coronary fistulas are less common, but usually drain into the right ventricle or right atrium. While coronary-

cameral fistulas are abnormal connections between a coronary artery and a cardiac chamber, the embryological basis of coronary artery to pulmonary artery fistulas can be based on Hackensellner's involution persistence hypothesis[1] proposing that there are six anlages in the truncus of which two ,that are seen in the aortic sinuses, persist and give rise to the coronary arteries while the others involute. Accordingly, the normally involuted anlage from the pulmonary sinus persists and connects with the anlage from the aortic sinus giving rise to these fistulas.[2] Small congenital CAF can be asymptomatic, larger more complicated CAFs can cause congestive heart failure, myocardial ischemia due to a coronary steal effect, thrombosis, rupture and infective endocarditis. Symptomatic CAF can be treated by selective ligature of the fistula tract or transcatheter closure.

CAF can be found in isolation, or as in the present case in association with other structural heart disease. The postnatal diagnosis can be suspected by echocardiography, but cardiac catheterization is required to precisely evaluate the vascular anatomy[3]. Isolated giant CAF has been diagnosed at 38 weeks of gestation, and successfully treated in the neonatal period[4]. The prenatal diagnosis of ventriculo-coronary artery fistula in the setting of PA-IVS has also been made previously[5]. In the literature coronary-pulmonary artery fistulas (CPAF) have been reported in association with TOF and pulmonary atresia in 10 % and the majority of them have a fistula originating from the left coronary artery with >50 % also having systemic-to-pulmonary collateral arteries (MAPCAs)[6].

Only one report described the prenatal diagnosis of a large fistula from the left coronary artery to the left atrial appendage in TOF, with congestive cardiac failure postnatally and death after surgical correction of TOF with ligation of the fistula at 3 months of age[7].

We did not find the association of TOF , CPAF and agenesis of the ductus arteriosus as described in the present case. The fistula was only diagnosed at autopsy , but prenatally we found a discrepancy between the narrow pulmonary annulus with a thickened pulmonary valve with minimal flow at 22 weeks and the pulmonary trunk that was adequately developed. This adequate pulmonary development can be caused by extra blood supply through the large fistula.

ETHICS

Approvement of the ethical commission was not required for this manuscript.

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Legend to the figures

Figure 1 Ultrasound with white pulmonary valve(arrow) , well developed trunk (dot)and right sided aorta.

Figure 2 Macroscopic view on the heart with  aorta,  pulmonary artery,  right coronary artery and  the fistula.